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CASE REPORT

Pediatric/Craniofacial

A Case of Pediatric Melanoma: Treatment Considerations in Advanced Disease

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Summary: We document a 3-year-old healthy African American girl who developed malignant melanoma on her lower extremity. The clinical appearance offered little indication of the lesion's severity (T4), and only the history of de novo presentation and disproportionate growth raised clinical suspicion. This case report highlights the subtle clinical findings of this condition and presents controversies related to surgical management of pediatric melanoma. *(Plast Reconstr Surg Glob Open 2015;3:e402; doi: 10.1097/GOX.0000000000000370; Published online 18 May 2015.)*

Each year, more than 76,000 new cases of melanoma are diagnosed in the United States.[1] Pediatric melanomas account for 0.7% of all cases and 0.1% of subsequent mortalities. The incidence of pediatric melanoma rises 2.6% annually, which may be partially explained by changes in sun-related behavior, yet the use of tanning beds and increases in ultraviolet exposure do not account for rises in prepubertal populations.[1–3] As the number of children diagnosed with melanoma continues to rise, accurate and timely diagnosis and treatment becomes even more paramount. Clinical suspicion remains the cornerstone of diagnosis and depends on an appreciation of the unique clinical features of pediatric melanoma.

Risk factors for melanoma common to both pediatric and adult populations include Fitzpatrick 1 skin phenotype, positive family history, previous malignancies, concomitant medical conditions such as xeroderma pigmentosum, and chronic immunosuppression.[3,4] In contrast to established risk factors for adults, sun exposure does not correlate with prepubertal, pediatric melanoma. Rates of disease are highest in areas of low ultraviolet-B exposure based on geographic location in that patient population.[1–3] The incidence of melanoma in adolescents, on the other hand, more closely mirrors the adult melanoma population correlation with increased ultraviolet-B exposure.

Traditional detection criteria used in adults often fail to identify malignant lesions in children. Suspicious clinical findings in children include amelanosis, bleeding, raised lesions, color uniformity, and de novo lesion development.[3,5] Furthermore, classic prognostic features, including tumor depth of invasion and lymph node involvement, less clearly impact survival in children.[6,7] We report on a case of melanoma in a 3-year-old patient to address the clinical features of this disease in the pediatric population and review some of the factors used to guide treatment options.

CLINICAL REPORT

A 3-year-old African American girl was referred to our office at the request of her dermatologist for a de novo, suspicious lower extremity lesion. On physical examination, she demonstrated a 0.6 × 0.5 cm, raised, darkly pigmented, well circumscribed anterior right knee lesion (Fig. 1). There was no ulceration, bleeding, or surrounding skin changes. The patient was healthy. She had no medical conditions and took no medications, and there was no personal or family history of skin cancer. An excisional biopsy was performed with wide local excision. Histologic evaluation demonstrated malignant melanoma, nevoid type, with a 4.95 mm tumor depth (T4) characterized

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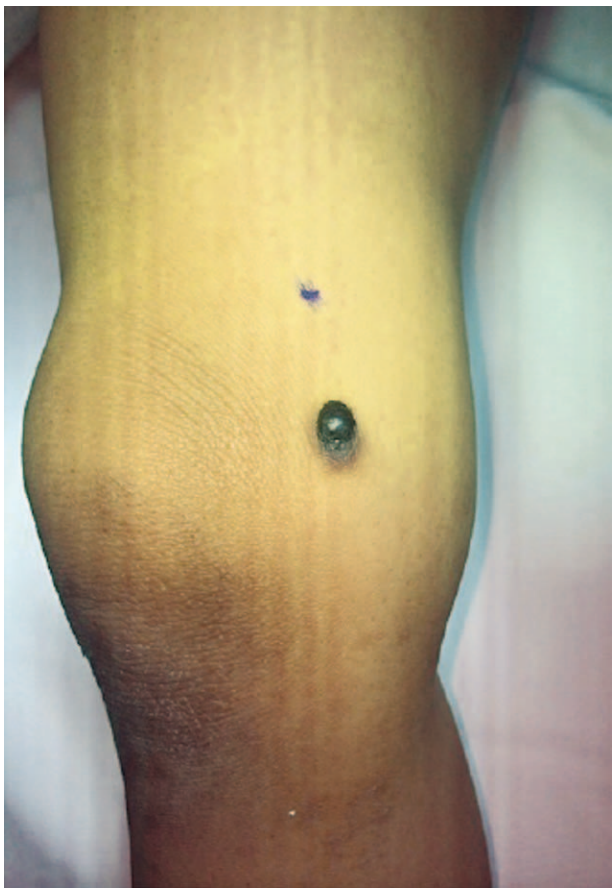

Fig. 1. Three-year-old girl with right anterior knee lesion.

by high mitotic activity, poor maturation, and junctional/dermal proliferation of atypical melanocytes (Fig. 2). Immunohistochemical staining established loss of p16 expression.

The patient returned to the operating room 3 weeks later for lymphoscintigraphy using 0.4 mCi of technetium 99m injected in 4 doses along the previous surgical scar. A single sentinel lymph node was identified and removed immediately preceding a longitudinal reexcision of the biopsy scar with a 2-cm margin and primary closure. Unfortunately, the sentinel node was positive for a focus of metastatic disease, and ultimately, the patient underwent radical groin dissection, which was negative for any residual nodal involvement.

DISCUSSION

In contrast to the established diagnostic, therapeutic, and prognostic criteria for adult melanoma, the clinical features of pediatric melanoma are poorly characterized. Traditional "*ABCD*" detection criteria must be expanded for pediatric screening to include amelanosis, color uniformity, and newly arising lesions.[5] Diagnosing pediatric melanoma is further complicated by histologic similarities with some forms

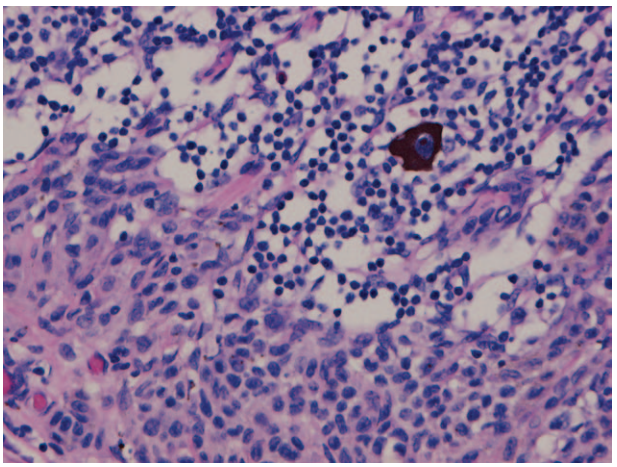

Fig. 2. Digital photograph of the lesion visualized with hematoxylin and eosin stain with high mitotic activity, poor cellular maturation, and atypical melanocytes (×20 magnification).

of benign melanocyte proliferation. Large congenital melanocytic nevi (CMN) remain a main risk factor for pediatric melanoma; patients with CMN have a 52% higher risk of melanoma compared with the general population. This risk is greatest for patients with large CMN of the trunk, lesions greater than 20 cm, and multiple satellite nevi.[8,9] Moreover, 16% of melanomas are initially misdiagnosed as Spitz nevi.[1,4] Lymph node biopsies have been advocated to aid in obtaining the correct diagnosis.[7] Gill et al[10] found that *B-RAF*, *N-RAS*, and *H-RAS* mutations are unique to spitzoid melanomas compared with the Spitz nevi.[1]

In the adult population, sentinel lymph node biopsy (SLNP) is an important staging procedure with prognostic implications.[6,7,11,12] The prognostic role of SLNB for pediatric melanoma has not yet been determined. Roaten et al[6] have demonstrated, however, that SLNB correlates with tumor depth and is safe without mortalities or recurrences over 35 months of follow-up. By contrast, the adult cohort of this study had fewer cases of sentinel lymph node involvement (18%) but overall fared worse with a 25% recurrence rate and 9% mortality rate within 19 months from their SLNB.[6] Whether there is a greater propensity for nodal metastasis, a delay in clinical diagnosis, or another explanation, remains to be determined; however, pediatric melanoma frequently involves local lymph node basins. The prognostic implications for a positive SLNB on recurrence and survival are either not supported or not yet established.[5–7,13] Once nodal involvement has been established, the need for further therapeutic intervention remains unclear. It is important to carefully discuss the known morbidity of completion lymph node dissection (CLND) in the context of its undetermined benefit. Fourteen percent of patients with a positive sentinel lymph node

will have residual tumor draining to their lymph node basin, which may be detected by CLND.[7] Complication rates following CLND approach 60%, including lymphedema (19–28%) and infection (17–28%).[7,13] As with SLNB, the benefit of CLND for pediatric melanoma is largely unknown. To extrapolate from adult data, however, 5-year survival rates are higher following immediate CLND (72.3%) in patients with node-positive disease compared with delayed lymphadenectomy when clinically apparent regional node metastasis develop (52.4%).[11]

Our patient with stage III melanoma and node-positive T4 tumor bears a 5-year survival rate estimated at 77%. Nevertheless, her age and the absence of tumor ulceration may prove more favorable characteristics that improve her prognosis.[4] The family elected to pursue a CLND despite a candid discussion about the minor risks involved with the procedure and the inconclusive benefits of the procedure. Her procedure went well and she is undergoing medical adjuvant treatment of her cancer. As additional cases of pediatric melanoma arise, long-term outcomes need to be evaluated to determine the appropriate surgical management of these tumors to most accurately counsel patients and their families.

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PATIENT CONSENT

Parents or guardians provided written consent for the use of the patient's image.

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